Epitomes

Important Advances in Clinical Medicine

Pediatrics

The Scientific Board of the California Medical Association presents the following inventory of items of progress in pediatrics. Each item, in the judgment of a panel of knowledgeable physicians, has recently become reasonably firmly established, both as to scientific fact and important clinical significance. The items are presented in simple epitome and an authoritative reference, both to the item itself and to the subject as a whole, is generally given for those who may be unfamiliar with a particular item. The purpose is to assist busy practitioners, students, research workers or scholars to stay abreast of these items of progress in pediatrics that have recently achieved a substantial degree of authoritative acceptance, whether in their own field of special interest or another.

The items of progress listed below were selected by the Advisory Panel to the Section on Pediatrics of the California Medical Association and the summaries were prepared under its direction.

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Kawasaki Disease

KAWASAKI DISEASE (mucocutaneous lymph node syndrome), originally known by pathologists as infantile periarteritis nodosa, began to be recognized as a clinical entity in Japan and in the United States in the late 1960s. It has now been seen in most countries of the world, but it remains a disease of unknown cause that is increasing in incidence and that has a propensity to affect young children, especially those of Japanese or part-Japanese racial extraction. Cases have been diagnosed in more than 50,000 children of all races, and an unknown number remain unidentified.

The major clinical findings, described separately by Dr Kawasaki in Japan in 1967 and by Dr Melish in the United States in 1974, are fever lasting longer than five days and averaging 11 days in duration; conjunctival injection of blood vessels; changes of the mouth consisting of erythema of the lips followed by fissuring and crusting, diffuse reddening of the oropharynx and prominent papillae of the tongue ("strawberry" tongue), and involvement of the extremities including swelling of the hands and feet, reddening of the palms and soles, erythematous rash and lymphadenopathy. Also seen to a lesser degree are pyuria and urethritis, arthralgia and arthritis, aseptic meningitis, vomiting, diarrhea, abdominal pain, myocardiopathy, pericardial effusion, obstructive jaundice, hydrops of the gallbladder, acute mitral insufficiency and myocardial infarction. In recent years there have been patients who have not shown full-blown disease. Physicians need a high degree of awareness, using the five major criteria to arrive at a diagnosis.

Most of the patients are children 2 years old or younger, or are younger than 8 years of age. Boys outnumber the girls 1.5:1 in clinically recognized cases. These infants and young children are generally very ill and very irritable. Because they do not respond to antipyretic therapy or antibiotics (or both), they tend to be seen by their private physicians frequently during the first week of illness and are admitted to hospital because of a combined lack of response to therapy and the development of other signs and symptoms such as a rash or joint involvement.

Pertinent laboratory findings include an elevated leukocyte count during the febrile phase of more than 20,000 per μl with a shift to the left. The erythrocyte sedimentation rate and C-reactive protein are increased from the febrile phase for six to ten weeks. Other positive results often are small numbers of leukocytes in cerebrospinal fluid and in urine specimens, slightly elevated liver function values and occasional jaundice. Thrombocytosis occurs in all patients between the 15th and 25th day of illness, with a peak platelet count of 600,000 to 1.8 million per μl. This correlates with the period of acute coronary artery thrombosis.

This is a clinical disorder that is similar to a number of other childhood illnesses and, for this reason, the diagnosis is made only after other diseases have been ruled out. While the disease is self-limited within six to eight weeks in most cases, about 1.7% of patients will die of cardiac complications, including coronary artery aneurysms and thrombosis (most commonly), atrioventricular conduction abnormalities or, rarely, cardiac tamponade. Peripheral vasculitis is widespread throughout the body and affects large and medium-

sized musculoelastic arteries and, to a lesser degree, veins. The ratio of male to female patients in fatal cases is 3:1, and most are younger than 2 years of age at the time of death.

Special tests are required to diagnose coronary artery aneurysms, which affect about 20% of patients. Two-dimensional echocardiography is sensitive in detecting left coronary artery aneurysms. Generally, both coronary arteries are aneurysmally dilated in the proximal area. Most of the coronary artery lesions regress in size within a year while the rest have persistent aneurysm or narrowed, tortuous arteries or at least minor permanent damage.

The treatment of Kawasaki disease is supportive care. Until recently administration of high doses of aspirin was the treatment of choice, but this is now undergoing further study. It seems likely that a lower dosage of aspirin will be recommended in the near future. Steroid therapy is contraindicated. In an unpublished study of 300 patients by Drs Kusakawa and Yanagawa of Japan, patients treated with steroid therapy had a significantly higher incidence of aneurysm formation than children treated only with aspirin. Patients with cardiac abnormalities are observed closely during and following their initial illness and should be seen periodically throughout childhood and adolescence for further cardiac or vascular complications of the illness.

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Lyme Disease

LYME DISEASE is caused by a spirochete that can be found in blood, skin lesion or cerebrospinal fluid specimens. It has also been isolated from the nymphal and adult forms of its vector, *Ixodes dammini*. In California and Oregon, *Ixodes pacificus* ticks have been implicated. Cases have been widely distributed over the United States, Europe and the Far East.

Lyme disease typically begins in the summer with a unique skin lesion, erythema chronicum migrans, which usually lasts about three weeks, beginning as a red macule or papule that expands to form a large ring with central clearing. Annular lesions usually range from 6 to 52 cm in diameter; they may be absent or number more than 20 and last from three days to eight weeks. The rash may be accompanied by fever, headache, stiff neck, myalgias, arthralgias, malaise, fatigue or moderate lymphadenopathy. Weeks or months later, aseptic meningitis, meningoencephalitis, cranial neuropathies especially of the facial nerve, myelitis, migratory musculoskeletal pain, monoarticular or oligoarticular arthritis, myocarditis or atrioventricular node block may develop in certain patients. Radiculoneuritis may occur that is indistinguishable from brachial plexus neuritis. A Guillain-Barré-like syndrome, atypical in that it may show a cerebrospinal fluid pleocytosis in the range of 35 to 120 leukocytes, with 60% to 100% as mononuclear cells, has been described. Lymphocytic meningoradiculitis (Bannwarth's syndrome) has been associated serologically with Lyme disease. Serum cryoprecipitates, raised serum IgM levels and elevated erythrocyte sedimentation rates may occur.

Penicillin or tetracycline given for ten days can successfully treat the early phases of the disease when rash is present and can prevent, or at least ameliorate, the subsequent arthritic, neurologic or cardiac disorders.

Diagnosis can be confirmed by the finding of raised levels of IgG or IgM antibodies to the spirochete using indirect immunofluorescence, or by isolating the spirochete. These tests are presently of limited availability except through state departments of health, so local laboratory personnel should be alerted to the specific diagnostic concern when considering this disease.

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Pneumococcal Polysaccharide Immunization

IMMUNIZATION WITH pneumococcal polysaccharide vaccine is a safe and effective means of increasing a specific antibody and providing protection against overwhelming infection from *Streptococcus pneumoniae*. It is generally agreed, however, that immunization should be restricted to specific populations at high risk for infection.

Those patients who should be immunized include persons with increased susceptibility to infection but in whom a normal antibody response can develop following immunization. Specific diagnoses include sickle cell disease, splenectomy following trauma or for hematologic disorders, aging (that is, older than 55 years of age), complement disorders and nephrotic syndrome. Patients who might benefit from immunization but who have an impaired antibody response include those who have Hodgkin's disease with splenectomy, multiple myeloma and other malignant disorders. Many patients who are susceptible to overwhelming infection have severely impaired immunity and would not be expected to benefit from immunization. These include patients who have hypogammaglobulinemia and severe abnormalities of both T-cell and B-cell immunity. Patients with chronic lung disease who do not have an increased susceptibility to *S pneumoniae* infection, such as those who have asthma or cystic fibrosis, should not be immunized.

Immunization should be given to children 2 years